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CASE REPORTS

Situs Inversus Dextrocardia with Cyanogen Complex Cardiopathy in a 16-Year-Old Albanian Male

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Abstract

Introduction: Dextrocardia is rare in the general population and may be associated with significant additional cardiac malformations. It is commonly associated with additional cardiac malformations.

In this report, we have described the follow-up of a patient with Situs inversus dextrocardia and cyanogen complex cardiopathy in a 16-year-old Albanian male. The male patient, born in 2007 in Albania, was referred to our ambulatory at six months of life by a pediatrician because of cyanosis and cardiac murmur. The echo Color Doppler examination was performed, with the conclusion: Situs inversus dextrocardia, unique ventricle, pulmonary arterial atresia. In 2008, a diagnostic catheterization was performed. The medico-chirurgical consultation has decided to leave the boy in natural history with a periodic follow–up. On 06.2009, in one of the routine examinations, there was evidence of hypertrophy of the unique ventricle associated with arterial hypertension. From that time, the patient is under medical treatment with periodic monitoring.

Conclusions: The regular follow-up of complex cyanogen congenital heart disease improves health care for the risqué target group.

In a heave, desaturation patients, hypertension must be evaluated as a secondary complication of the primary problem.

Keywords: dextrocardia; hypertension; situs inversus; unique ventricle.

Introduction

Dextrocardia is rare in the general population and may be associated with significant additional cardiac malformations. [1] In all its presentations, dextrocardia is a rare congenital abnormality, and while its true incidence remains unknown mainly, estimates range from 1 in 8,000 to 25,000 live births.[2] It is commonly associated with additional cardiac malformations, the frequency of which varies considerably according to Situs: 5% with situs inversus to 90% with situs solitus.[3, 4, 5] No predilection for race, ethnicity, or gender has been described for dextrocardia. Dextrocardia has an estimated incidence of around 1 in 12,000 pregnancies and may be associated with other cardiac anomalies.[6]

Dextrocardia can occur with an average position of abdominal visceral organs (situs solitus), with a reversal in the position of abdominal visceral organs (situs inversus), or with the abnormal distribution of major abdominal visceral organs.[7] In this report, we described the follow-up of a patient of 16-year-old Albanian male presenting Situs inversus dextrocardia with cyanogen complex cardiopathy

Case Report

The male patient was born in 2007 in Albania, from a normal pregnancy, at term, birth weight: Kg 2,800, and was diagnosed at two months old with congenital heart disease. The patient was referred to our ambulatory on 30.04.08 by a pediatrician because of a lack of weight gain and growth in height, sweating, cyanosis, and cardiac murmur. In the objective examination, it was observed that the saturation in basal conditions, in ambient temperature, was 75-80%,

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with discrete stature and weight growth, with generalized muscular hypotrophy, cyanosis, symmetric thorax, and vesicular respiration.

An objective examination of the heart found dextrocardia, a strong systolic and diastolic murmur audible at all points, and the murmur covered T1 and T2.

An objective examination of the abdomen: is palpable, liver at 1 cm to the left of the costal arch. Clinical examinations show:

Chest x-ray: There is no evidence of pleuro-parenchymal leisure; pulmonary hilus shadow and vascular branch are accentuated. Hart's shadow is positioned on the right, in an atypical shape, and has significantly increased. The peri-broncho-vascular interstitium is increased. ECG: Sinus rhythm, Fc 140/min, poor left ventricular presence, right atrial overload. ECHO Color Doppler: dextrocardia, situs inversus. Normal pulsative Abdominal Aorta. The inferior and superior vena cava are drained into the anatomical right ventricle positioned on the left. The pulmonary vein drained in the anatomical left ventricle, positioned on the right. Huge interatrial defect. There is no connection from the left atrium to the left ventricle. Only one atrioventricular valve, probably tricuspid connected with the ventricle, is anatomically right-positioned on the left. It is visualized as a rudimentary chamber on the right, like the left ventricle, in the position anteriorly on the right in front of the dominant ventricle. This rudimentary chamber connects the dominant ventricle with a restrictive Ventricle Septal Defect. Trivial tricuspid insufficiency. From the unique ventricle originates the aorta in a posterior position. The pulmonary artery or pulmonary trunk is not seen. Two small pulmonary branches, the left pulmonary branch 3mm and the right pulmonary branch 2mm, take flow only from the Botally duct. (See figure nr1 A.)

Conclusion: Dextrocardia, Situs atrial, inversus + Single Atrium + Single Ventricle + Mitral Valve atresia, transposition of the great arteries, pulmonary atresia with confluent branches, bilateral systemic-pulmonary collateral circles.

So, on 06.2008, a diagnostic Cardiac catheterization was performed in a specialized center for congenital heart diseases in Italy.

After medical-chirurgical collegial discussions at the same diagnostic center, it was decided to leave the child in natural history with periodic follow-up because of the problematic anatomical correction.

In June 2009, a routine examination with cardiac ultrasound revealed evidence of hypertrophy of the unique ventricle associated with arterial hypertension. In Figure 1 A, we present the first documentation of hypertrophy of the unique ventricle caused by arterial hypertension. The hypertension was symmetrically 130/80 mmHg in both arms, and the saturation of O_2 was 75% at ambient temperature. Face edema and declined bilateral edema were present.

Based on the primary diagnosis, the following examinations were requested: ECG, RX, Renal ultrasound, renal blood tests (azotemia 69 mg/dl, uricemia 8.1mg/dl, creatinine clearance 25.75ml/min, proteinuria of 24 h was 4350mg, diuresis 500ml/24h). In the context of chronic hypoxia, suspecting renal failure, a consultation with a pediatric nephrologist was requested. After this, he was under treatment with Amlodipine 5mg 1X1tab, Enalapril 25 mg 2X1/2 tab, Allopurinol 300 mg 1X3/4 tab, Furosemide 40 mg (1/2-0-0) tab. Atenolol 100 mg 1X1/4 tab. By regularly taking antihypertensive medicine, the restoration of the blood balance for azotemia and uricemia was constant; arterial pressure was under control and saturation of O_2=88%.

For the monitoring of hypertension and the therapeutic antihypertensive effect, in addition to recurrent arterial pressure measurements, we also used echocardiography, measuring the thickness of the wall of the unique ventricle. In Figure 1 B on 07/03/2012, we presented the cardiac ultrasound image. (See Figure 1. B)

On 07/2022, echocardiography showed a remodeled cardiac unique ventricular hypertrophy. (Figure 2 A) Blood proper brachial pressure 110/70mmHg and left

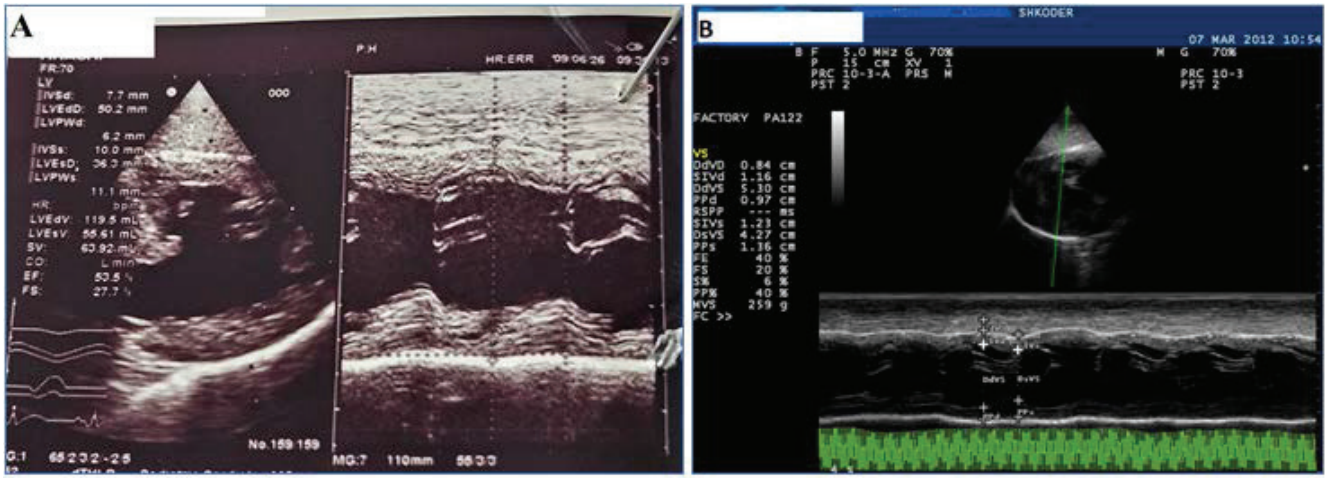

Figure 1. (A on 26/06/2009 first echocardiography, B echocardiography on 07/03/2012)

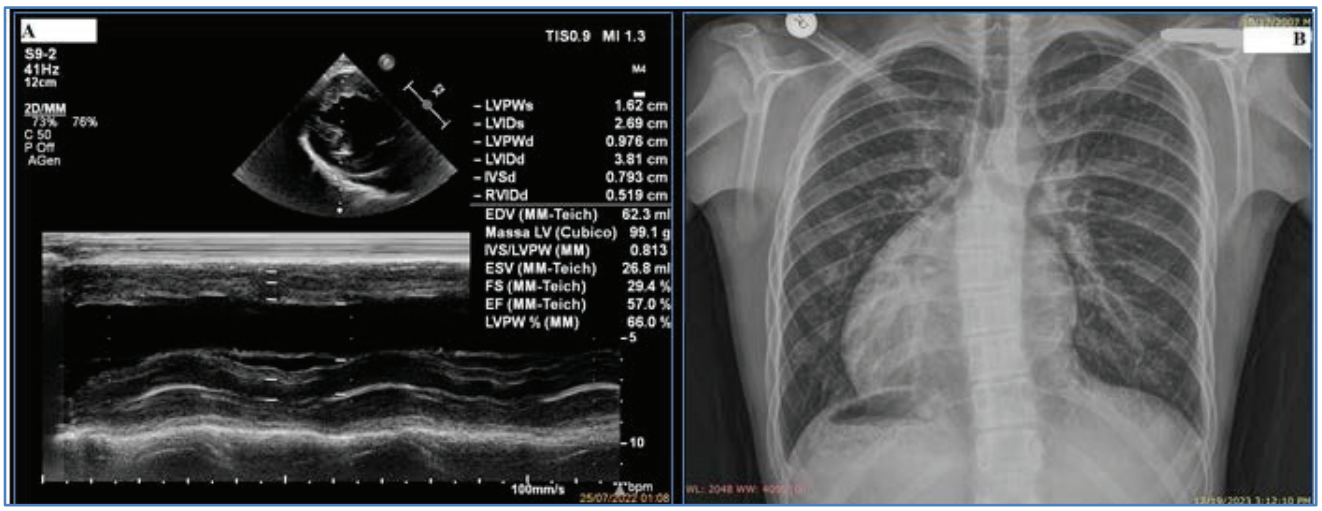

Figure 2. (A. echocardiography on 25/07/2022; B. Chest x-ray on 19/12/2023)

105/70mmHg. Chest x-ray on 12/2023 (See figure 2 B): apex cordis on the right site raised above the diaphragm, dextrocardia, cardiomegaly, cardiac, thoracic index 0.64, prominence of II cardiac arch on the right, prominence of I cardiac arch on the left, low evidence of the pulmonary vascular tree, gastric bubble on the right site, liver on the left site saturation of O_2=81%.

The 48-hour blood pressure Holter report on 28/04/2024 evidences a stable situation. The blood pressure is within the normal limit values. The therapy with antihypertensive drugs dominates very well the blood pressure during physical activities. It is recommended to continue the same schematic therapy.

Figure 3 shows that the average Systolic blood pressure is between 90 and 130 mmHg, and diastolic blood pressure is between 40 and 60 mmHg.

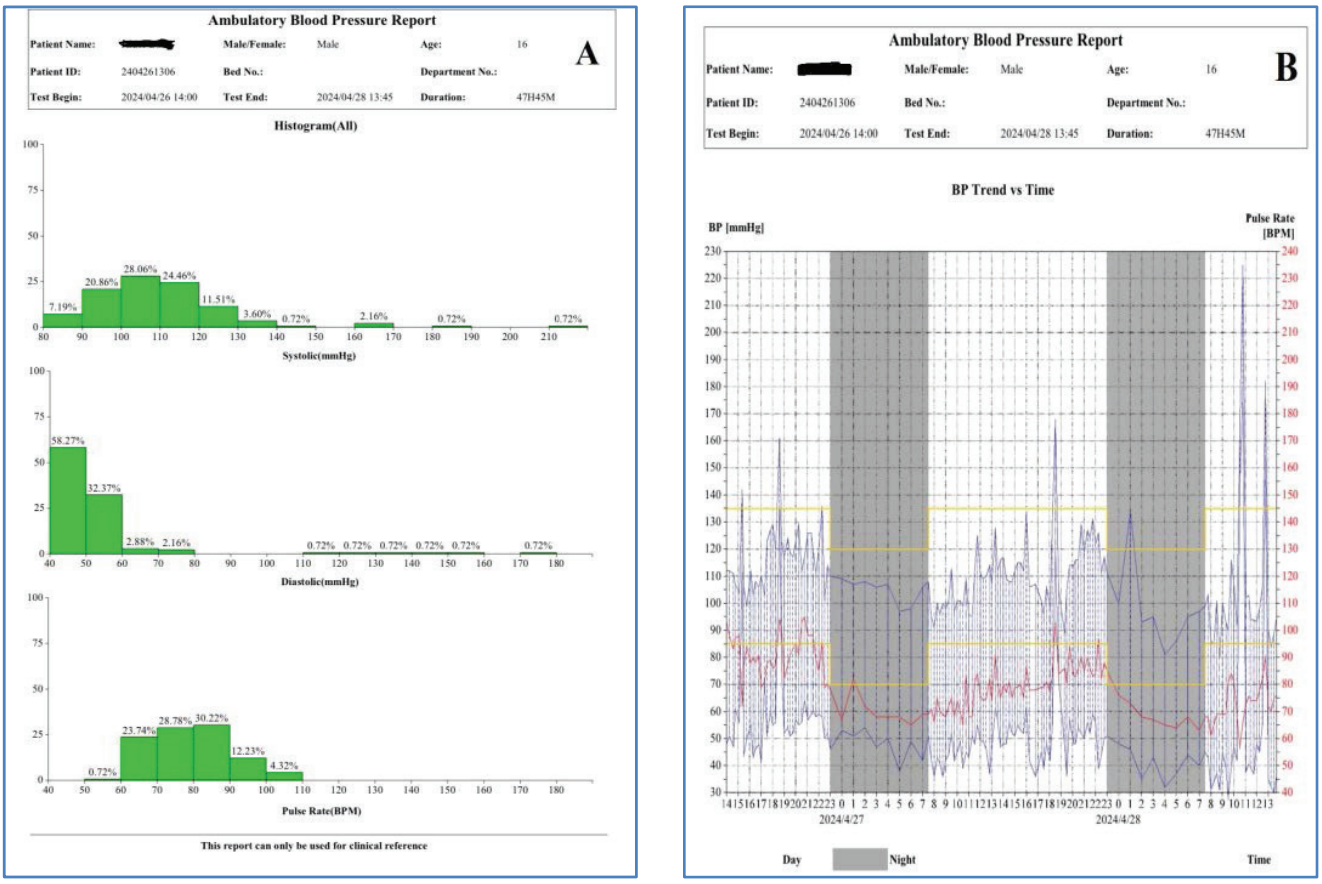

Figure 3. A&B Holter pressure data results

Conclusions

The regular follow-up of complex cyanogen congenital heart disease improves health care for the risqué target group.

In a heave, desaturation patients, hypertension must be evaluated as a secondary complication of the primary problem.

Declaration of Conflicting Interests
The Authors declare that there is no conflict of interest.

Founding
This research received a specific grant from the University of Shkodra, "Luigj Guarakuqi."

Ethical Consideration
Ethical approval for this study has been reviewed and approved by the Ethical Committee, Faculty of Natural Sciences, Department of Clinical Subjects, University of Shkodra No. Prot. 87. Dt. 09/05/2024, Shkodër Albania

Statement Consent
The patient provided written informed consent for the publication and any accompanying images. The editor-in-chief of this journal can review a copy of the written consent upon request.

Ethics Approval
Ethical approval for this study has been reviewed and approved by the Ethical Committee, Faculty of Natural Sciences, Department of Clinical Subjects, University of Shkodra No. Prot. 87. Dt. 09/05/2024, Shkodër Albania

Human and Animal Rights
No animals were used in this research. All procedures performed in studies involving human participants were according to the standards of institutional and research committees and the 1975 Declaration of Helsinki, as revised in 2013.

Availability of Data and Materials
The data supporting the findings of the article is available by request from the primary author, [APG].

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Author Contribution Statements
APG - writing the paper and providing the radiological imagery and interpretations; ZSh- collecting the data, writing the paper, drafting the article, and revising it; EP- revising the article.

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